A FURTHER CONTRIBUTION TO THE PATHOLOGY OF ARRESTED CEREBRAL DEVELOPMENT.

By B. SACHS, M.D.

THE gross cerebral lesions of childhood have received much attention of late years, and the conditions which lead to the development of paralysis of epilepsy and of idiocy, or possibly of all three conditions combined, are tolerably well understood. Among the large number of cases of this description that I have had occasion to examine, one small group has attracted my special attention. It includes possibly the severest form of idiocy (the palsy or epilepsy, if present, being of secondary importance). It runs in families, a number of children of the same family being similarly afflicted, the disease running the same course in each case; these children appear to do well until about the fifth to eighth month; then a marked retrograde-movement sets in, all the cerebral functions, sensory and motor, become impaired, the child becomes idiotic, blind, more or less paretic, and fortunately dies, promptly enough, of marasmus.

The first case of this description, I reported to this Association five years ago. I may claim for this case that it stands as one of the first in which a minute histological examination of the cortex had revealed the changes underlying the development of one form of idiocy.

The patient, who is the chief subject of this paper, is a sister of the child whose brain I reported upon five years ago. A healthy child, a boy, has been born between these two children, and is an exceptionally bright child. The occurrences of this same form of idiocy in

---

[1] Read at the meeting of the American Neurological Association, New York, June 22, 23 and 24, 1892.

several members of one family I have observed a number of times.

Last August, a lady brought to me her boy, R. H., aged thirteen months. This was her tenth child, five children having died; of these, three were exactly like the one brought to me, and all died before the age of two years of general marasmus. The mother has had no miscarriages, and I could not reasonably entertain even a suspicion of syphilis, the mother also denied a neurotic family taint; she was an exceptionally healthy looking woman, and all her children were entirely normal after easy labors. The child brought to me is said to have weighed twelve pounds at birth; was nursed at the breast, and did well until six or seven months old; then began to droop its head; had its first teeth at seven months; since that time child has been steadily losing, mentally and physically; the fontanelles were still open, the head moderate in size; there was a general spastic condition in both legs, and is not able to, nor does not attempt to, walk; has only the barest perception of light, but is excessively startled by every sound; the eyes diverge, but there is no nystagmus; it has occasional slight spasms, but no convulsive seizures. I have not heard of the child since, but I have no doubt it has joined the preceding three.

The prompt development of blindness with idiocy in all these cases—I have the history of six such cases—points to what we find—a morbid process involving equally frontal, parietal and occipital portions of the cortex.

The history of the child whose entire central nerve axis has been subjected to careful histological examination is as follows:

The child was thirteen months old at the time of my first consultation with Dr. S. B. Jones. The experiences with the first child led the parents to exercise unusual care in raising this child; it was born at full term, and was of entirely normal physical development; was given the bottle at first, later on was nursed at the breast; it did fairly well, and appeared to be as far advanced as any child until the age of eight months; since that time there has been steady retrogression. On closer examination it was found that the child could not hold its head up, nor could it sit up unless supported; it has no perception of light; the pupillary light reflex is entirely gone; it has slight sense of hearing, but does not distinguish between sounds; there is a slight spastic condition of upper and lower extremities; no spontaneous movement of any sort; child sits listlessly on nurse's lap; the knee-jerks on both sides were distinctly exaggerated; no ankle clonus was obtainable; plantar reflexes were also increased; at this time one tooth was breaking through; the fontanelles were not entirely closed. I entered upon my notes at the time: Resemblance to first child very close in every respect except that the paresis is more spastic in character than in the first child. The effect of treatment was, of course, *nil.*

Six months later I had an opportunity of seeing the child again, together with Dr. Kimball. The child had gone down hill steadily; sight and hearing were completely lost; the mind was entirely blank, and the child died at the age of twenty months (four months earlier than the first child) of an extreme marasmus. A week before death had considerable fever and several convulsive seizures; but these were the only ones that had occurred during the entire course of the disease. I performed the autopsy twenty-two hours after death. In order to determine whether there was latent specific disease, I made a most careful examination of all the organs of the body, and of the larger glands, but I found no proof of any such taint; the liver, heart, lungs, spleen, and intestines were entirely normal, except that all the organs were remarkably pale; the heart and aorta were of normal size. The skull was symmetrical, of good size, but unusually thin; the fontanelles were closed, but still transparent. The dura was adherent to the skull so that the brain had to be removed with the calavarium. As in the case of child No. 1, the brain was firm to the touch, almost as hard. I noted at the time, as a brain that has been in Müller's fluid for some weeks, the pia could be easily detached, and the cortex, though pale, presented no unusual appearances.

The anomalies of fissuration, such as were noted in the first case, were not present here.

The spinal cord presented normal macroscopical appearances. The brain and spinal cord were at once immersed in Müller's fluid and carefully hardened. Careful sections were made of all parts of the cortex of the ganglia, of the optic chiasm, of the pons medulla, and of the lower portion of the cord. The cervical and dorsal portions of the cord were, unfortunately, lost in some mysterious way.

All the newer stains were employed by Dr. Powers who assisted me in this part of the work, but very few and unimportant changes were found either in the ganglia, pons, medulla or cord; the chief changes, and very marked ones in this case as in the preceding ones, were found in the cortex, and in all regions of the cortex to an equal degree. I would have but to reproduce the drawings accompanying my first paper on this subject to illustrate and explain the specimens from this case. It is evident that the chief morbid condition is as represented by the cortical cells.

There is not in all the specimens I have examined a single normal pyramidal cell; in all the layers the cell bodies are altered either in shape or in general appearances; the cell nucleus and the maleolus are distinct enough, as a rule, but surrounded by an altered cell body which does not properly take the ordinary stains. The neuroglia cells appear somewhat increased, and the tissue rather dense. The blood-vessels are present in at least the usual proportion, most of them filled with blood, but no sign of any active inflammatory condition. I attached special importance to the determination of this point, as a paucity of blood supply might have been supposed to have been the starting-point of this trouble.

The white fibres entering the cortex showed in departure from the normal, as can be seen on numerable Weigert specimens, but I have not succeeded in demonstrating tangential fibres. The ganglia were carefully examined, but their cellular structure was found normal. The cord was but imperfectly examined, for reasons stated above, as the specimens taken from the lower segments, it was evident that the cellular development was quite perfect, and there was no distinct degeneration in the white columns.

Comparing the results of this examination with the facts published a few years ago, that this arrest of cerebral development is practically an arrest of *cortical* development—a true *agenesis corticalis*, but by what immediate forces this is brought about it is difficult to say. In the first case the history of traumatism during pregnancy was worked as a possible cause, but in the present instance this factor is wanting. We must fall back upon the neurotic taint; in the case of the other children mentioned (four in one family) there is no proof of even a "neurotic taint." We are therefore somewhat at sea as regards the ætiology of this special form of idiocy, but on the strength of the two autopsies now placed on record to which we might add similar findings of Kast and Jensen, we may claim the chief morbid changes underlying that form of idiocy which is associated with blindness and which leads to a prompt, fatal termination. The absence of epilepsy is worthy of note. This can hardly be dignified as a distinct *type* of diseases, but it represents the most pronounced form of idiocy and of arrested cerebral development.